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Extra Skeletal Manifestations of Histocytosis in Pediatrics

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Abstract : Background: Langerhans cell histiocytosis (LCH) is a rare multi-systemic disease that shows an abnormal proliferation of these kinds of cells associated with a granular infiltration that affects different structures of the human body, including the lung, liver, spleen, lymph nodes, brain, mucocutaneous, soft tissue (head and neck), and salivary glands. Evaluation of the extent of disease is one of the major predictors of patient outcome. Objectives: To recognize the pathogenesis of Langerhans cell histiocytosis (LCH), describe the radiologic criteria that are suggestive of LCH in different organs rather than the bones and to illustrate the appropriate differential diagnoses for LCH in each of the common extra-osseous sites. Material and methods: A retrospective study was done on 150 biopsy-proven LCH patients from 2007 to 2012. All patients underwent imaging studies, mostly US, CT, and MRI. These patients were reviewed to assess the extra-skeletal manifestations of LCH. Results: In 150 patients with biopsy-proven LCH, There were 33 patients with liver affection, 5 patients with splenic lesions, 55 patients with enlarged lymph nodes, 9 patient with CNS disease and 11 patients with lung involvement. Conclusions: Because of the frequent LCH children and evaluation of the extent of disease is one of the major predictors of patient outcome. Radiologist need to be familiar with its presentation in different organs and regions of body outside the commonest site of affection (bones). A high-index suspicion should be raised a biopsy is recommended in the presence of radiological suspicion. Chemotherapy is the preferred therapeutic modality.

Keywords : langerhans cell histiocytosis, extra-skeletal, pediatrics, radiology
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